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IMAGING CASE STUDY OF THE MONTH

ISOLATED UNILATERAL FIBROUS DYSPLASIA OF THE SPHENOID SINUS

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INTRODUCTION

Fibrous dysplasia as a clinical term was introduced by Lichtenstein[1] as early as 1938. The disease is characterized by a slowly progressive replacement of cancellous bone by abnormal fibrous tissue that histologically represents various stages of bone metaplasia.[2] The proportions of the fibrous and osseous tissues vary, occasionally even in the same bone.[3,4] The development of the disease lasts for years and presents with deformity or mild symptoms. This locally destructive disease can occur in association with endocrine abnormalities such as Albright's syndrome (precocious puberty, cutaneous pigmented patches over the involved bones) or as an isolated clinical entity. Rarely, fibrous dysplasia is associated with other bone pathological changes such as aneurysmal bone cyst inside the paranasal sinuses[5] or mucocele.[6] The cause of the disease, however, remains unknown.

Ramsey et al[7] offered a classification of fibrous dysplasia into 3 types: type 1, characterized by single or multiple lesions in 1 and the same bone (monostotic type); type 2, characterized by multiple lesions involving more than 1 bone (polyostotic); and type 3, characterized by dissemination of pathological bone changes connected to other disturbances such as Albright's syndrome. There are also classifications according to histologic and radiologic findings.[8,9] In terms of the predisposing sites of its clinical appearance, craniofacial fibrous dysplasia is most frequently located in the mandible and maxilla.[2,10-12] The paranasal sinuses are rarely involved. The most common locations are the maxillary and, less frequently, ethmoidal sinuses.[13] To the best of our knowledge, an isolated, unilateral fibrous dysplasia of the sphenoid sinus has not been reported so far in the literature.

CASE REPORT

A 21-year-old man with a history of recent onset of severe headaches was evaluated by computed tomography (CT) and magnetic resonance imaging (MRI). An undefined "mass" was found within the remarkably enlarged right sphenoid sinus (Figs 1 and 2). To exclude a possible relationship between the mass and the internal carotid artery, as well as the right cavernous sinus, arteriography of both external and internal carotid arteries was performed, and the results were negative. An endoscopic surgical approach to the right sphenoid sinus was performed.

The posterior third of the middle turbinate had to be removed to make the anterior sphenoid wall more visible. However, despite the relatively good visi-

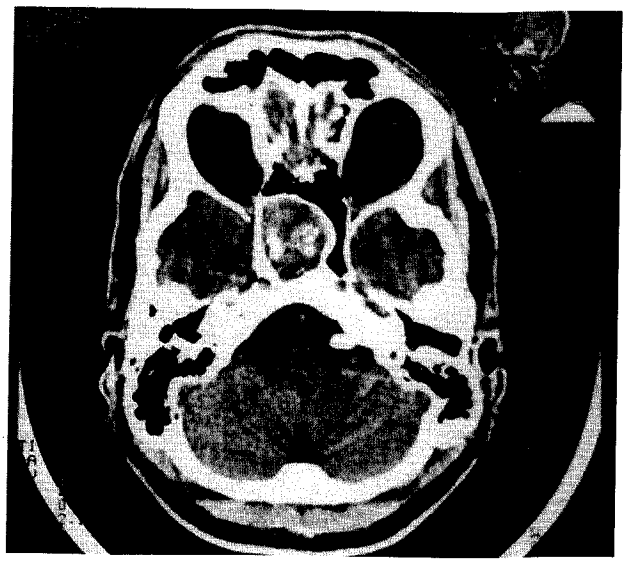

Fig 1. Enhanced axial computed tomogram of sphenoid sinus showing sclerosis and emphasized expansion of right sphenoid sinus. Surrounding bony structures seem to be of normal appearance. Computed tomography appearance of sinus content is characterized by heterogeneous density (ground-glass appearance). Isolated, bone-like particles of irregular, bizarre shape inside involved sinus not only closely resembled osteosarcoma's spicules, but also suggested aspergilloma.

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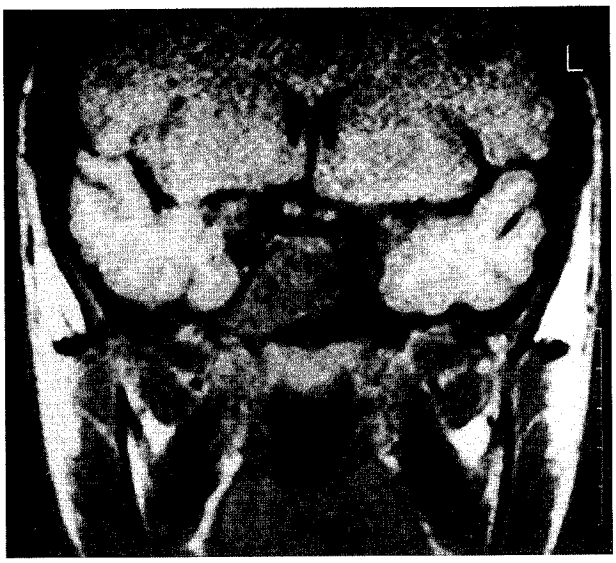

Fig 2. Magnetic resonance coronal image showing bizarre formation of heterogeneous density in projection of right sphenoid sinus. No signal characteristics suggesting calcification can be seen within mass.

bility and all surgical endeavours, the crucial maneuver of opening the anterior wall by a direct approach through the sphenoethmoid recess remained unsuccessful. Hence, an additional, more classic endoscopic approach to the sphenoid sinus through the ostiomeatal complex and posterior ethmoid was performed. This helped ensure that the site of previous attempts in opening the sphenoid sinus was correct. Finally, it became obvious that we were dealing with "bone in bone": after the partial removal of an unexpected bony "shell," an unusual bony-fibrous tissue was found behind it. The tissue was very adherent to the adjacent sinus walls, making removal, particularly in the zones of the optic nerve and carotid artery, very tricky. Nevertheless, a vast majority of the suspect tissue was removed. Histopathologic analysis of the frozen sections revealed fibrous dysplasia. Proliferative, well-vascularized fibrous stroma surrounding the irregularly shaped trabeculae of woven bone without osteoblastic activity was found. Its appearance was intraluminal, suggesting the origin of this unusual mass to be from the sphenoid sinus septum (Fig 3). Fifteen months after the surgery, the patient is doing well.

DISCUSSION

Fibrous dysplasia develops mostly during the first 2 decades of life and usually regresses at puberty, although it may continue to grow slowly in adulthood.[14,15] It seems that this was the case in our patient.

Fibrous dysplasia in an "active phase," ie, prior to its presumed regression in adolescence, can show a very dense vascularity. The increased vascularity of active lesions may cause symptomatic arteriovenous shunting.[16] Fortunately, we did not find any arteriovenous shunting during arteriography of the internal and external carotid arteries. Further, some authors state that even in symptomatic patients with biopsy-proved fibrous dysplasia, because of a dense vascularity the surgery should be delayed when feasible, until the growth of fibrous dysplasia slows or ceases.[17]

With craniofacial involvement, visual loss may be caused by compression of the optic nerve[18] or chiasm[19] by dysplastic bone, or by hemorrhage into dysplastic bone.[20] In our patient we did not find any clinical, CT, or MRI signs of involvement of other structures, except the right sphenoid sinus. To the best of our knowledge, this is the first report of an isolated, unilateral fibrous dysplasia of the sphenoid

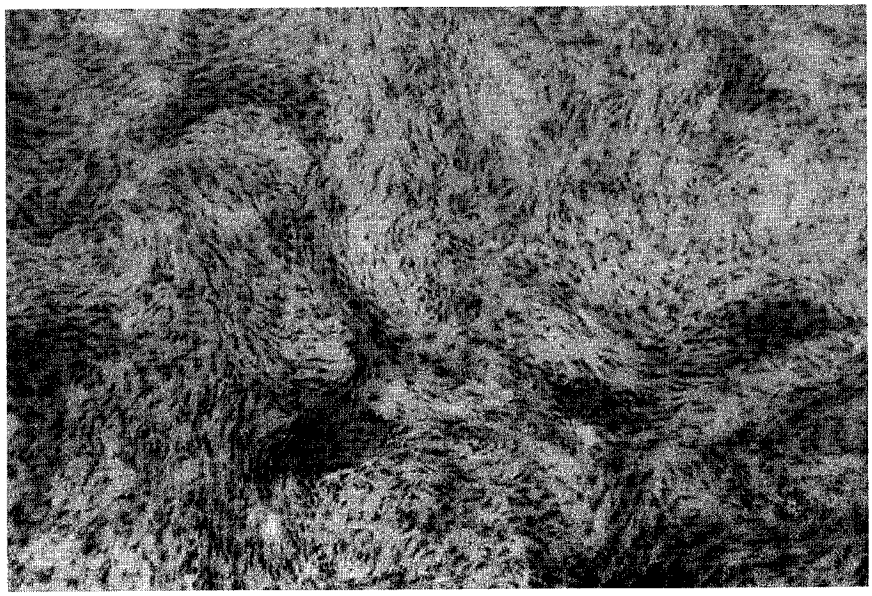

Fig 3. Proliferative, well-vascularized fibrous stroma surrounding irregularly shaped trabeculae of woven bone without osteoblastic activity (H & E, original ×100).

sinus. The most similar case was that described by Mueller et al[10] in 1992, but the disease was not entirely isolated, since they found the ipsilateral great wing of the sphenoid bone to be involved as well.

Concerning the differential diagnosis prior to surgery, it must be emphasized that radiologic signs, although they can be very characteristic, are not always easy to recognize. Fries[8] has classified craniofacial fibrous dysplasia into 3 radiographic patterns: pagetoid, sclerotic, and cystlike. The pagetoid form resembles Paget's disease, with remarkable bone expansion and alternating zones of radiodensity and radiolucency. This type more frequently appears in patients over 30 years of age, and the symptoms can last for more than 15 years on average. The sclerotic type demonstrates a homogeneous, severe density, again accompanied by bone expansion, whereas the cystlike pattern is usually a single, well-defined radiolucent lesion, round or oval in shape, edged by a thin, sclerotic margin clinically resembling an eggshell. Both are frequently seen in children or younger adults (less than 25 years of age) who have had symptoms for less than 3 years.

Fries'[8] classification, however, was established on the basis of conventional radiography. Computed tomography diagnosis has introduced the term "ground glass–like lesion." It usually measures 70 to 130 Hounsfield units (HU), contrary to other lesions, such as osteomyelitis and neoplasms, which usually have lower attenuations of 20 to 40 HU.[9] The bonelike spicules, characteristic of osteosarcomatous lesions, must be differentiated from aspergilloma, since aspergilloma may also present with sporadic opacities of almost bony density.[21] In contrast to osteosarcomatous lesions, these opacities are mostly located in the center of the lesion.

On MRI, fibrous dysplasia displays nonspecific findings, with low signal intensity on T1-weighted images and low, intermediate, or high signal intensity on T2-weighted images.[22-24] It seems that fibrous dysplasia may not be differentiated on the basis of signal intensity alone, ie, by means of standard MRI (Fig 2). This technique may not demonstrate a thin calcific shell and calcifications as effectively as CT (Fig 1).[16] Our patient showed many "characteristic" radiologic and MRI signs, which confused us.

From the histopathologic point of view, woven bone usually predominates in typical fibrous dysplasia, but various amounts of lamellar bone may also be seen.[16] The microscopic differential diagnosis of fibro-osseous clinical entities of the facial bones includes fibrous dysplasia, ossifying fibroma, and cementifying fibroma.[14] They all resemble each other, so it is difficult to distinguish among them. Still, ossifying fibromas are often well delimited and are composed of lamellar bone, lined by osteoblasts. Cementifying fibromas are made up of spherules with interspersed fibroblastic changes; the spherules are interpreted as cementum.

The histology in our case showed an ill-defined lesion made up of proliferating fibroblasts in a compact stroma of interlacing collagen fibers. Irregular C-shaped or Chinese character–shaped trabeculae of the woven bone were scattered throughout the lesion. Osteoblastic rimming was not observed (Fig 3).

In regard to secondary malignant degeneration of fibrous dysplasia, pain and soft tissue extension should be strongly considered, since sarcomatous alterations occur in 0.5% of all patients.[25,26] More than half of these sarcomas are located in the craniofacial region. Osteosarcoma is the most common secondary tumor in fibrous dysplasia.[23]

Finally, there is the problem of how to adequately follow up patients operated on for fibrous dysplasia, since it is difficult to distinguish among recurrent lesions, postoperative and postirradiation inflammation, and secondary sarcomatous degeneration. It seems that a new MRI technique, gadolinium-enhanced dynamic subtraction MRI, may be useful because it can separate the rapid, early vascular phase of enhancement of malignant tumors from the slower, late vascular phase of enhancement of benign lesions and inflammations.[27]

In conclusion, fibrous dysplasia of the sphenoid sinus presents a diagnostic challenge to both clinicians and radiologists. It must be considered in cases of a calcified, thick margin of the enlarged sphenoid sinus and a ground-glass appearance of the mass inside it. Because of the potential for malignant change, vigilant clinical and imaging assessment of fibrous dysplasia with histopathologic correlation is essential.

REFERENCES

1. Lichtenstein L. Polyostotic fibrous dysplasia. Arch Surg 1938;36:874-7.

2. Antonelli JR. Ossifying fibroma of the maxillary sinus. A case report. Ann Dent 1989;48:33-6.

3. Kiehn CL, Desprez JD, Harris AH. Fibrous dysplasia of the facial bones. Am J Surg 1961;102:835-7.

4. Smith JF. Fibrous dysplasia of the jaws. Arch Otolaryngol 1965;81:592-603.

5. Som PM, Schatz CJ, Flaum EG, Lauman TH. Aneurysmal bone cysts of the paranasal sinuses associated with fibrous dysplasia: CT and MR findings. J Comput Assist Tomogr 1991; 15:513-5.

6. Garzozi H, Garty J, Kavch Z. Craniofacial fibrous dysplasia complicated by mucocele. The role of radionuclide scinti-

graphic methods in the diagnosis. Ann Ophthalmol 1989;21: 108-10.

7. Ramsey HE, Strong EW, Frazel EL. Fibrous dysplasia of the craniofacial bones. Am J Surg 1968;116:542-7.

8. Fries YW. The roentgen features of fibrous dysplasia of the skull and facial bones: a critical analysis of 39 pathologically proved cases. AJR Am J Roentgenol 1957;77:71-8.

9. Daffner RH, Kirks DR, Gehweiler JA Jr, Heaston DK. Computed tomography of fibrous dysplasia. AJR 1982;139:943-7.

10. Mueller DP, Dolan KD, Yuh WTC. Fibrous dysplasia of the sphenoid sinus. Ann Otol Rhinol Laryngol 1992;101:100-1.

11. Rifai M, Kenawi B. Visual dysfunction: abnormal sign of benign sphenoethmoidal lesions. J Laryngol Otol 1990;104: 217-21.

12. Barat M. Fibrous dysplasia masquerading as chronic maxillary sinusitis. Ear Nose Throat J 1989;42:44-8.

13. Dornhoffer J, Schwager K. Fibrous dysplasia and ossifying fibroma. Two unusual fibro-osseous lesions of the paranasal sinuses. HNO 1995;43:193-6.

14. Fechner RE. Problematic lesions of the craniofacial bones. Am J Surg Pathol 1989;13(suppl 1):117-32.

15. Henry A. Monostotic fibrous dysplasia. J Bone Joint Surg Br 1969;5B:300-6.

16. Shapeero LG, Vanel D, Ackerman L, et al. Aggressive fibrous dysplasia of the maxillary sinus. Skeletal Radiol 1993; 22:563-8.

17. Harrison DFN. Osseous and fibro-osseous conditions affecting the craniofacial bones. Ann Otol Rhinol Laryngol 1984; 93:199-203.

18. Ogusthorpe JD, Gudeman SK. Orbital complications of fibrous dysplasia of the skull. Otolaryngol Head Neck Surg 1987;97:403-5.

19. De Fanti C, Rivier A. Syndrome chiasmatique sur dysplasie fibreuse du sphenoide. Rev Otoneuroophthalmol 1972;44: 339-46.

20. Kurokawa Y, Sohma T, Tsuchita H, Kitami K, Suzuki S, Sohma K. Hemorrhage into fibrous dysplasia following minor head injury — effective decompression for the ophthalmic artery and optic nerve. Surg Neurol 1989;32:421-6.

21. Stammberger H, Jakse R, Beaufort F. Aspergillosis of the paranasal sinuses. X-ray diagnosis, histopathology, and clinical aspects. Ann Otol Rhinol Laryngol 1984;93:251-6.

22. Norris MA, Kaplan PA, Pathria M, Greenway G. Fibrous dysplasia: magnetic resonance imaging appearance at 1.5 Tesla. Clin Imaging 1990;14:211-4.

23. Som P, Bergeron RT. Head and neck imaging. St Louis, Mo: Mosby–Year Book, 1991:109.

24. Utz JA, Kransdorf MJ, Jelinek JSS, Mosser RP Jr, Berrey BH. MR appearance of fibrous dysplasia. J Comput Assist Tomogr 1989;13:845-9.

25. Schwartz DT, Alpert M. Malignant transformation in fibrous dysplasia. Am J Med Sci 1964;247:1-20.

26. Bagatin M, Manojlovic S, Zajc I, Cacinovic-Matic V. Monostotic fibrous dysplasia of the facial bones. Acta Stomatol Croat 1993;27:51-7.

27. Shapeero LG, Vanel D, De Baere T, Valenty M. Gadolinium-enhanced subtraction MRI: new technique for evaluating response of osteosarcoma to chemotherapy. Radiology 1990; 181:227-33.